## Fetiform Teratoma (Homunculus)

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25-year-old woman (gravida 1, para 1) presented with pelvic pain. A pelvic ultrasound demonstrated  $\sim$  a 12 cm imes 13 cm complex mass in the right adnexa, suggesting a large dermoid cyst. Preoperative serum human chorionic gonadotropin (hCG) was negative. She underwent a laparotomy and right ovarian cystectomy. The ovarian cyst was as described below:

Ovarian cyst, right: Received fresh, the specimen consists of a 220-gram partially encysted soft tissue mass that extrudes abundant hair and sebaceous debris. The cyst is opened to reveal a fetus-shaped fragment of soft tissue, 7 cm in cephalic/caudal length . . . . Two upper limbs are identified, 2 cm and 1.5 cm in maximum length. The longer has 1 finger, 0.8 cm. Two lower limbs are identified. One measuring 3.2 cm in length has 3 grossly recognizable toes, 1 of which has a toenail. The other limb, measuring 3.5 cm in length, has 2 grossly recognizable toes, 1 with a grossly recognizable nail (arrow). The entire surface of the specimen is hair bearing and is covered with yellow sebaceous material. The specimen is opened with a Y-shaped incision to reveal a tubular structure, grossly consistent with bowel. . . .

Karyotype: mos 47,XX,+7/46,XX

Final Diagnosis: Fetiform teratoma (homunculus), 220 grams

Fetiform teratoma (homunculus) is a term that has been given to a rare form of teratoma that resembles a malformed fetus. There are very few reported cases of this entity in the English-language literature. This tumor must be distinguished from both fetus in fetu (a parasitic monozygotic twin usually found inside the body of a newborn or infant) and an ectopic pregnancy.

Mature cystic teratomas are common benign ovarian tumors that occur most frequently in women of reproductive age. The majority are composed of disorganized, neoplastic, mature tissues of 1 or more of the embryonic germ layers: ectoderm, mesoderm, and endoderm. Rarely, these tumors develop a high degree of differentiation and organization, resembling a malformed fetus (fetiform structure). It has been proposed that fetiform teratoma and fetus in fetu can be differentiated based on zygosity. Most ovarian

teratomas are homozygous at loci where the host normal tissue demonstrates heterozygosity, but fetus in fetu is genetically identical to its host.

Fetiform teratomas must also be distinguished from ectopic pregnancies. All reported cases of fetiform teratoma are composed of mature tissue and present without placental or trophoblastic tissue. A clinical history of an elevated  $\beta$ -hCG level and the documentation of chorionic tissue can substantiate the diagnosis of an ectopic pregnancy.<sup>1</sup>

## Reference

 Weiss JR, Burgess JR, Kaplan KJ. Fetiform teratoma (homunculus). Arch Pathol Lab Med. 2006;130:1552-1556.

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